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Case of Mediastinal Ewing's Sarcoma/Primitive Neuroectodermal Tumor Presenting as Pleural Effusion

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I. Introduction

Ewing sarcoma is a small round blue cell tumour with regular sized primitive appearing cells. It is closely related to the soft tissue tumours pPNET, Askin tumour and neuroepithelioma, which collectively are referred to as Ewing sarcoma family of tumours (ESFT) . They share not only microscopic appearances but also demonstrate a non-random t(11;22)(q24;q12) chromosome rearrangement.

II. Material

A young male aged 22yrpresented with complaints offever, breathlessness,dry cough,left sided chest pain since 1 month to JJM Medical College. Patient was subjected to Chest x ray and CECT(16 slice TOSHIBA machine) and blood investigations.

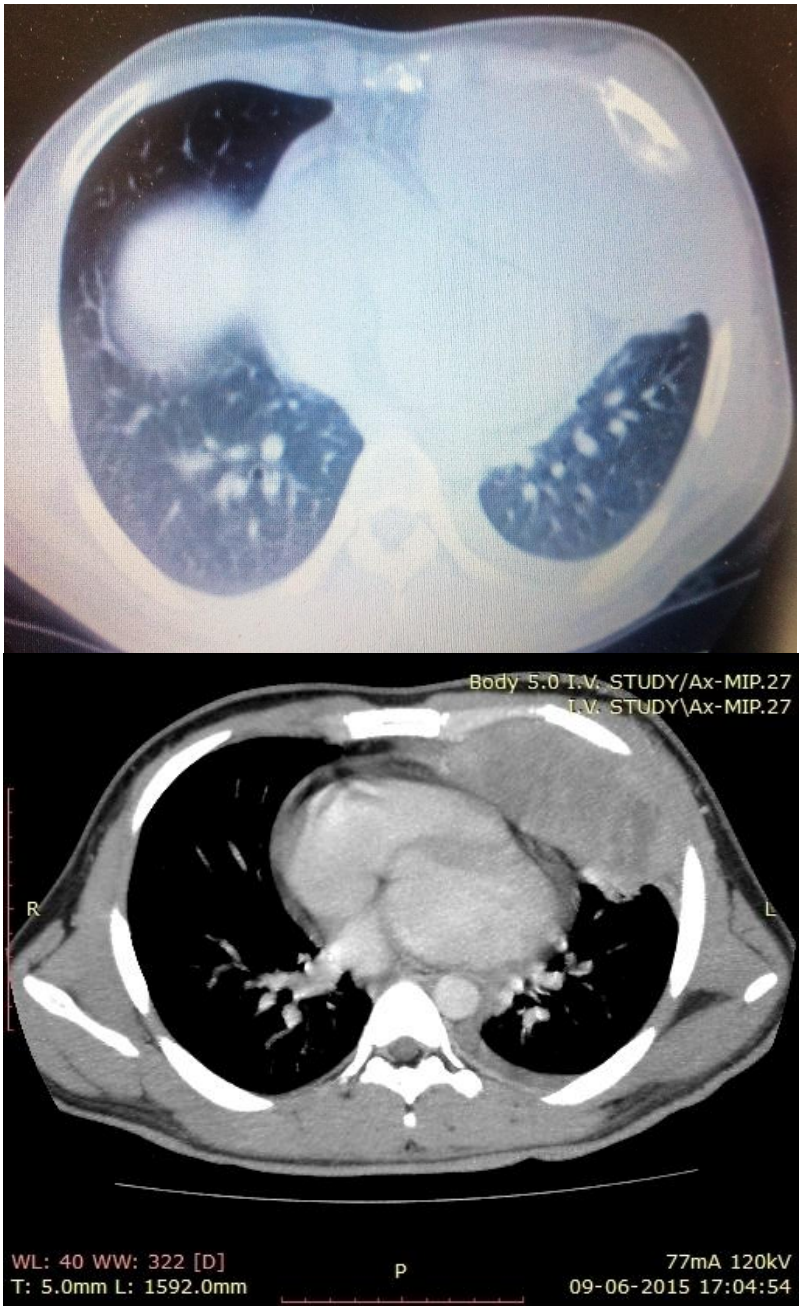
III. .Examination

On physical examination vitals were stable, Signs of left pleural effusionwithtrachea shiftedto rightside. Blood Investigations shows Complete Blood Count,Renal FunctionTest,LiverFunctionTest- Within Normal Limit , sputum forA F B-negative.CXR-Signs of moderate pleural effusion, mediatinum shifted to theright side;Pleural fluid analysis:straw-colored,[TC– 170/cumm, L- 80%, N-15%, ADA-122 IU, protein-3.7g/dl, sugar- 86 mg/dl] **malignant cells –negative**.

CECT THORAX

CT machine used was 16 slice TOSHIBA machine

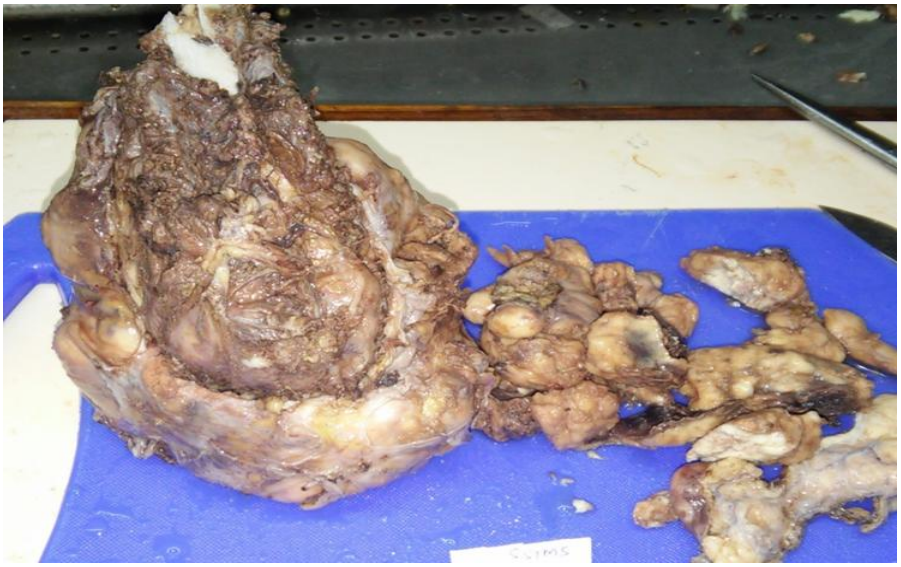
CT findings- Well-defined heterogeneous soft tissue density mass lesion noted predominantly within the anterior mediastinum measuring 15x13x10 cm making obtuseangles withanterolateral thechest wall causing medial displacement of the pleural and erosion of the left 5th ribwith minimal pleural effusion.lesion shows significant enhancement on contrast study.**F/S/O Ewing's Sarcoma of chest wall/ primitive neuroectodermal tumor (PNET).**Other differentials include **chondrosarcoma (**malignant tumour of the rib seen in elderly age group with calcification) and **osteosarcoma**;less likely pleural based tumour as pleura is displaced medially.



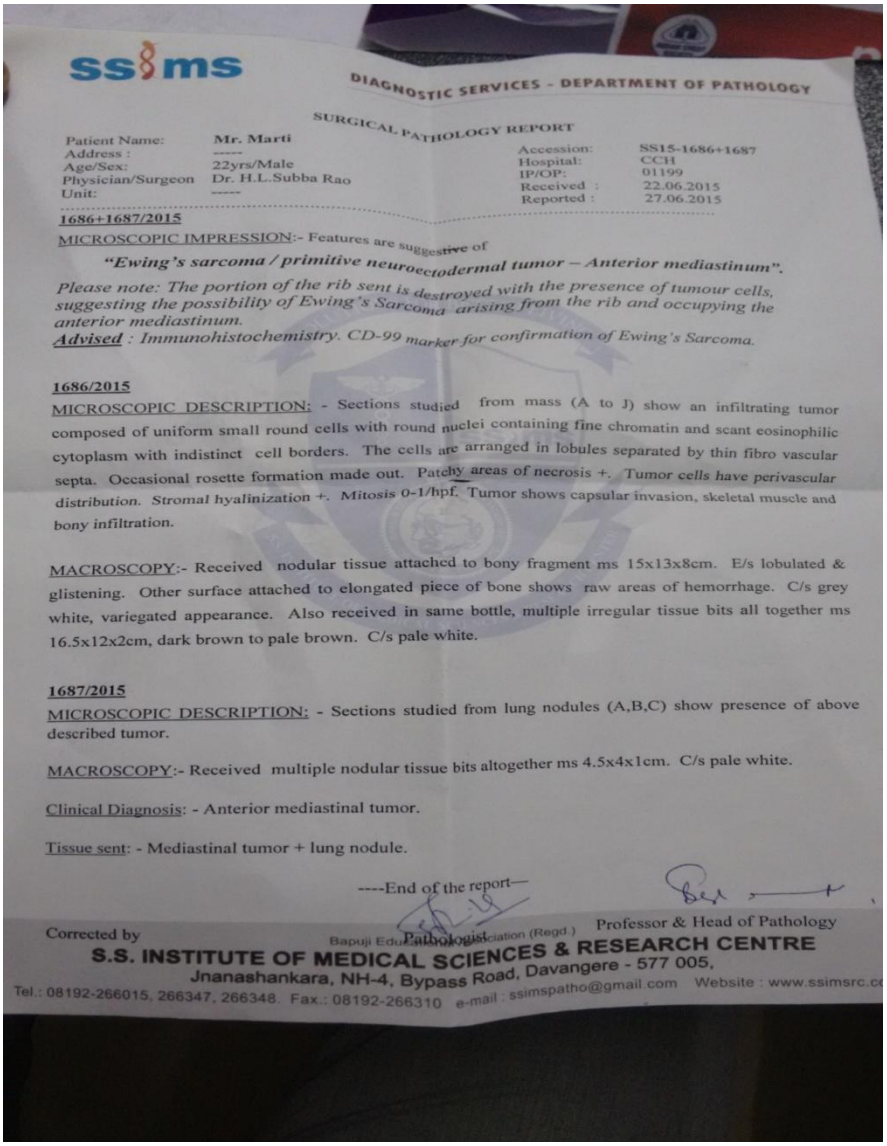
Axial CECT images showing :Well-definedheterogeneoussoft tissue density mass noted measuring15x13x8cmmaking obtuseangles withanterolateral chest wall causing medial displacement of the pleural and erosion of 5th ribwith minimal pleural effusion.
USG GUIDED **FNAC**:**F/S/OpPNET/Askin tumor**.

IV. Management

Patient underwent surgery tumor was excised completely from anterior chest wall and subjected to Histopathology examination of mass lesion shows: Small round cells with round nuclie containing fine chromatin and scant eosinophilic cytoplasm with indistinct cell borders. The cells are arranged in lobules separated by thin fibrovascular septa. Occasional rosette formation made out. Tumor shows capsular invasion, skeletal muscle and bony infiltration **F/S/O Ewing's sarcoma /primitive neuroectodermal tumor**.



Post operative specimen



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DIAGNOSTIC SERVICES - DEPARTMENT OF PATHOLOGY

SURGICAL PATHOLOGY REPORT

Patient Name:	Mr. Marti	Accession:	SS15-1686+1687
Address :	-----	Hospital:	CCH
Age/Sex:	22yrs/Male	IP/OP:	01199
Physician/Surgeon	Dr. H.L.Subba Rao	Received :	22.06.2015
Unit:	-----	Reported :	27.06.2015

1686+1687/2015

MICROSCOPIC IMPRESSION:- Features are suggestive of

"Ewing's sarcoma / primitive neuroectodermal tumor – Anterior mediastinum".

Please note: The portion of the rib sent is destroyed with the presence of tumour cells, suggesting the possibility of Ewing's Sarcoma arising from the rib and occupying the anterior mediastinum.

***Advised** : Immunohistochemistry. CD-99 marker for confirmation of Ewing's Sarcoma.*

1686/2015

MICROSCOPIC DESCRIPTION: - Sections studied from mass (A to J) show an infiltrating tumor composed of uniform small round cells with round nuclei containing fine chromatin and scant eosinophilic cytoplasm with indistinct cell borders. The cells are arranged in lobules separated by thin fibro vascular septa. Occasional rosette formation made out. Patchy areas of necrosis +. Tumor cells have perivascular distribution. Stromal hyalinization +. Mitosis 0-1/hpf. Tumor shows capsular invasion, skeletal muscle and bony infiltration.

MACROSCOPY:- Received nodular tissue attached to bony fragment ms 15x13x8cm. E/s lobulated & glistening. Other surface attached to elongated piece of bone shows raw areas of hemorrhage. C/s grey white, variegated appearance. Also received in same bottle, multiple irregular tissue bits all together ms 16.5x12x2cm, dark brown to pale brown. C/s pale white.

1687/2015

MICROSCOPIC DESCRIPTION: - Sections studied from lung nodules (A,B,C) show presence of above described tumor.

MACROSCOPY:- Received multiple nodular tissue bits altogether ms 4.5x4x1cm. C/s pale white.

Clinical Diagnosis: - Anterior mediastinal tumor.

Tissue sent: - Mediastinal tumor + lung nodule.

----End of the report----

Corrected by Pathologist Professor & Head of Pathology

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V. Observation

Ayoung male patient presented with fever, cough with a left pleural effusion. CECTthorax revealed mass lesion, patient underwent USG guided FNAC report S/O PNET. Patient underwent surgery, anterior mediastinal mass arising from chest wall resected with rib fragments subjected to HPEreported as "Ewing's sarcoma/primitive neuroectodermal tumor-Anterior mediastinum" Thedifferential diagnosis for chest wall tumor are

Benign		**Malignant**	
Soft tissue	**Skeletal (rib cage)**	**Soft tissue**	**Skeletal (rib cage)**
Haemangioma	Fibrous dysplasia (MC)	Rhabdomyosarcoma(MC)	Chest wall metastases(MC)
Lymphangioma	Aneurysmal bone cyst	Ewing'sarcoma;including Askin tumor/pPNET	Myeloma
Lipoma	Giant cell tumor	Ganglioneuroblastoma	Chondrosarcoma
Schwannoma	Ossifying fibromyxoid tumour	Neuroblastoma	Osteosarcoma
Neurofibroma	Osteochondroma	Angiosarcoma	
Ganglioneuroma	Chondromyxoid fibroma	Leiomyosarcoma	
Paraganglioma	Mesenchymal hamartoma of chest wall	Malignant fibrous histiocytoma	

VI. Conclusion

Ewing's sarcoma is highly malignant primarytumor. The tumor is derived from red bone marrow.most frequently,it is observed in children and adolescents aged 4-15 years and rarelydevelops in adults older than 30 years. Ewing sarcoma is the second most malignant tumor in young patients and it is the most lethal bone tumor. Males are affected than females.

Most frequently,the tumor is diagnosed as a monostotic lesion in the metaphysis or diaphysis of the long bones of the extremities. The tumor also may occur,although less frequently ,in the pelvic area,ribs,and scapulae.

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